Complete Summary

GUIDELINE TITLE

Left-sided heart obstructive lesions: aortic valve disease, subvalvular and supravalvular aortic stenosis, associated disorders of the ascending aorta, and coarctation. In: ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease).

BIBLIOGRAPHIC SOURCE(S)

Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, del Nido P, Fasules JW, Graham TP, Hijazi ZM, Hunt SA, King ME, Landzberg MJ, Miner PD, Radford MJ, Walsh EP, Webb GD, Smith SC Jr, Jacobs AK, Adams CD, Anderson JL, Antman EM, Buller CD, Creager MA, Ettinger SM, Halperin JL, Hunt SA, Krumholz HM, Kushner FG, Lytle BW, Nishimura RA, Page RL, Riegel B, Tarkington LG, Yancy CW. Left-sided heart obstructive lesions: aortic valve disease, subvalvular and supravalvular aortic stenosis, associated disorders of the ascending aorta, and coarctation. In: ACC/AHA 2008 guidelines for the management of adults [trunc]. J Am Coll Cardiol 2008;52(23):e185-97.

GUIDELINE STATUS

This is the current release of the guideline.

The guidelines will be reviewed annually by the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines and considered current unless they are updated, revised, or withdrawn from distribution.

COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

- Adult congenital heart disease
- Left-sided heart obstruction lesions, including:
    - Aortic valve disease
    - Subvalvular aortic stenosis
    - Supravalvular aortic stenosis
    - Aortic coarctation
    - Bicuspid aortic valve (BAV) abnormality

GUIDELINE CATEGORY

Counseling
Diagnosis
Evaluation
Management
Risk Assessment
Treatment

CLINICAL SPECIALTY

Cardiology
Family Practice
Internal Medicine
Radiology
Thoracic Surgery

INTENDED USERS

Health Care Providers
Physicians

GUIDELINE OBJECTIVE(S)

- To assist healthcare providers in clinical decision making by describing a range of generally acceptable approaches for diagnosis, management, and prevention of specific diseases or conditions associated with adult congenital heart disease (ACHD)
- To define practices that meet the needs of most patients in most circumstances
- To support the practicing cardiologist in the care of ACHD patients by providing a consensus document that outlines the most important diagnostic and management strategies and indicates when referral to a highly specialized center is appropriate

TARGET POPULATION

Adolescents, young adults, and older adults with congenital heart disease and left-sided heart obstructive lesions, including aortic valve disease, subvalvular and

supravalvular aortic stenosis, associated disorders of the ascending aorta, and aortic coarctation

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis/Evaluation

1. Imaging and hemodynamic assessment of aortic stenosis (AS)
2. Echocardiography
3. Cardiac catheterization
4. Coronary angiography
5. Electrocardiography
6. Magnetic resonance angiography
7. Exercise stress testing
8. Dobutamine stress testing
9. Transthoracic echocardiography (TTE)
10. Transesophageal echocardiography (TEE)
11. Brachial and femoral pulse palpation in aortic coarctation

Management/Treatment

Medical Therapy

1. Treatment of systemic hypertension (beta-blockers, angiotensin-converting enzyme inhibitors, or angiotensin receptor blockers)
2. Long-term vasodilator therapy for hypertension
3. Statins to prevent atherosclerosis

Surgical Interventions

1. Aortic balloon valvotomy
2. Aortic valvuloplasty, valve repair, or Ross repair
3. Surgical resection for subaortic stenosis
4. Surgical repair of supravalvular aortic stenosis
5. Interventions for coronary artery obstruction
6. Percutaneous catheter intervention for coarctation
7. Stent placement for long-segment coarctation
8. Surgical repair of coarctation

Follow-up

1. Lifelong cardiology follow-up
2. Reproductive counseling

MAJOR OUTCOMES CONSIDERED

- Progression or recurrence of aortic stenosis, regurgitation, or enlargement
- Complications of surgical interventions
- Mortality

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Hand-searches of Published Literature (Primary Sources)
Hand-searches of Published Literature (Secondary Sources)
Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Unlike other American College of Cardiology/American Heart Association (ACC/AHA) practice guidelines; there is not a large body of peer-reviewed published evidence to support most recommendations, which will be clearly indicated in the text. An extensive literature survey was conducted that led to the incorporation of 647 references. Searches were limited to studies, reviews, and other evidence conducted in human subjects and published in English. Key search words included but were not limited to adult congenital heart disease (ACHD), atrial septal defect, arterial switch operation, bradycardia, cardiac catheterization, cardiac reoperation, coarctation, coronary artery abnormalities, cyanotic congenital heart disease, Doppler-echocardiography, d-transposition of the great arteries, Ebstein's anomaly, Eisenmenger physiology, familial, heart defect, medical therapy, patent ductus arteriosus, physical activity, pregnancy, psychosocial, pulmonary arterial hypertension, right heart obstruction, supravalvular pulmonary stenosis, surgical therapy, tachyarrhythmia, tachycardia, tetralogy of Fallot, transplantation, tricuspid atresia, and Wolff-Parkinson-White. Additionally, the writing committee reviewed documents related to the subject matter previously published by the ACC and AHA. References selected and published in this document are representative and not all-inclusive.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Applying Classification of Recommendations and Level of Evidence

	SIZE OF TREATMENT EFFEC		
	CLASS I *Benefit >>> Risk* Procedure/Treatment	**CLASS IIa** *Benefit >> Risk* *Additional studies with focused objectives needed*	**CLASS IIb** *Benefit ≥ Risk* *Additional stud* *objectives nee* *registry data w*

		SIZE OF TREATMENT EFFEC		
		SHOULD be performed/administered	**IT IS REASONABLE** to perform procedure/administer treatment	*helpful* Procedure/Trea **MAY BE CONS**
Estimate of Certainty (Precision) of Treatment Effect	**LEVEL A** Multiple population evaluated* Data derived from multiple randomized clinical trials or meta-analyses	• Recommendation that procedure or treatment is useful/effective • Sufficient evidence from multiple randomized trials or meta-analyses	• Recommendation in favor of treatment of procedure being useful/effective • Some conflicting evidence from multiple randomized trials or meta-analyses	• Recomm usefuln less we • Greater evidenc multiple trials or analyse
	LEVEL B Limited population evaluated* Data derived from a single randomized clinical trial or nonrandomized studies	• Recommendation that procedure or treatment is useful/effective • Evidence from single randomized trial or nonrandomized studies	• Recommendation in favor of treatment of procedure being useful/effective • Some conflicting evidence from single randomized trial or nonrandomized studies	• Recomm usefuln less we • Greater evidenc random nonran studies
	LEVEL C Very limited population evaluated* Only consensus opinion of experts, case studies or standard of care.	• Recommendation that procedure or treatment is useful/effective • Only expert opinion, case studies, or standard-of-care	• Recommendation in favor of treatment of procedure being useful/effective • Only diverging expert opinion, case studies, or standard-of-care	• Recomm usefuln less we • Only di opinion or stan

*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as gender, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use. A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Even though randomized trials are not available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

Note: In 2003, the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines developed a list of suggested phrases to use when writing recommendations. All guideline recommendations have been written in full sentences that express a complete thought, such that a recommendation, even if separated and presented apart from the rest of the document (including headings above sets of recommendations), would still convey the full intent of the recommendation. It is hoped that this will increase readers' comprehension of the guidelines and will allow queries at the individual recommendation level. (See Table 1 in the original guideline document for a list of suggested phrases for writing recommendations.)

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

The committee reviewed and ranked evidence supporting current recommendations with the weight of evidence ranked as Level A if the data were derived from multiple randomized clinical trials involving a large number of individuals. The committee ranked available evidence as Level B when data were derived from a limited number of trials involving a comparatively small number of patients or from well-designed data analyses of nonrandomized studies or observational data registries. Evidence was ranked as Level C when the consensus of experts was the primary source of the recommendation. In the narrative portions of these guidelines, evidence is generally presented in chronological order of development. Studies are identified as observational, randomized, prospective, or retrospective. The committee emphasizes that for certain conditions for which no other therapy is available, the indications are based on expert consensus and years of clinical experience and are thus well supported, even though the evidence was ranked as Level C. An analogous example is the use of penicillin in pneumococcal pneumonia where there are no randomized trials and only clinical experience. When indications at Level C are supported by historical clinical data, appropriate references (e.g., case reports and clinical reviews) are cited if available. When Level C indications are based strictly on committee consensus, no references are cited. The final recommendations for indications for a diagnostic procedure, a particular therapy, or an intervention in adult congenital heart disease (ACHD) patients summarize both clinical evidence and expert opinion. The schema for classification of recommendations and level of evidence illustrates how the grading system provides an estimate of the size of treatment effect and an estimate of the certainty of the treatment effect (see "Rating Scheme for the Strength of the Evidence" above).

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

The American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines was formed to create clinical practice guidelines for select cardiovascular conditions with important implications for public health. This guideline writing committee was assembled to adjudicate the evidence and construct recommendations regarding the diagnosis and treatment of adult

congenital heart disease (ACHD). Writing committee members were selected with attention to ACHD subspecialties, broad geographic representation, and involvement in academic medicine and clinical practice. The writing committee included representatives of the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons.

Writing committees are specifically charged to perform a formal literature review, weigh the strength of evidence for or against particular treatments or procedures, and include estimates of expected health outcomes where data exist. Patient-specific modifiers, comorbidities, and issues of patient preference that might influence the choice of tests or therapies are considered, as well as the frequency of follow-up and cost-effectiveness. When available, information from studies on cost is considered, but data on efficacy and clinical outcomes constitute the primary basis for recommendations in these guidelines.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

See "Rating Scheme for the Strength of the Evidence" field, above.

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

External Peer Review
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

This document was reviewed by 3 external reviewers nominated from both the American College of Cardiology (ACC) and the American Heart Association (AHA), as well as reviewers from the American Society of Echocardiography, Canadian Cardiovascular Society, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, and Society of Thoracic Surgeons, and 20 individual content reviewers which included reviewers from the ACC Congenital Heart Disease and Pediatric Cardiology Committee and the AHA Congenital Cardiac Defects Committee. All reviewer relationships with industry information were collected and distributed to the writing committee and are published in the original guideline document (see the "Conflicts of Interest/Financial Disclosures" field in this document).

This document was approved for publication by the governing bodies of the American College of Cardiology Foundation (ACCF) and the AHA and endorsed by the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

The American College of Cardiology/American Heart Association (ACC/AHA) classification of the recommendations for patient evaluation and treatment (classes I-III) and the levels of evidence (A-C) are defined at the end of the "Major Recommendations" field.

Recommendations for Evaluation of the Unoperated Patient

Class I

1. Primary imaging and hemodynamic assessment of aortic stenosis (AS) and aortic valve disease are recommended by echocardiography-Doppler to evaluate the presence and severity of AS or aortic regurgitation (AR); left ventricular (LV) size, function, and mass; and dimensions and anatomy of the ascending aorta and associated lesions. (***Level of Evidence: B***)
2. Echocardiography is recommended for reevaluation of patients with AS who experience a change in signs or symptoms and for assessment of changes in AS hemodynamics during pregnancy. (***Level of Evidence: B***)
3. In asymptomatic adolescents and young adults, echocardiography-Doppler is recommended yearly for AS with a mean Doppler gradient greater than 30 mm Hg or peak instantaneous gradient greater than 50 mm Hg and every 2 years for patients with lesser gradients. (***Level of Evidence: C***)
4. Cardiac catheterization is recommended when noninvasive measurements are inconclusive or discordant with clinical signs. (***Level of Evidence: C***)
5. Coronary angiography is recommended before aortic valve surgery for coronary angiography in adults at risk for coronary artery disease. (***Level of Evidence: B***)
6. Coronary angiography is recommended before a Ross procedure if noninvasive imaging of the coronary arteries is inadequate. (***Level of Evidence: C***)
7. A yearly electrocardiogram (ECG) is recommended in young adults less than 30 years of age with mean Doppler gradients greater than 30 mm Hg or peak Doppler gradients greater than 50 mm Hg. (***Level of Evidence: C***)
8. An ECG is recommended every other year in young adults less than 30 years of age with mean Doppler gradients less than 30 mm Hg or peak Doppler gradients less than 50 mm Hg. (***Level of Evidence: C***)

Class IIa

1. In asymptomatic young adults less than 30 years of age, exercise stress testing is reasonable to determine exercise capability, symptoms, and blood pressure response. (***Level of Evidence: C***)
2. Exercise stress testing is reasonable for patients with a mean Doppler gradient greater than 30 mm Hg or peak Doppler gradient greater than 50 mm Hg if the patient is interested in athletic participation or if clinical findings differ from noninvasive measurements. (***Level of Evidence: C***)
3. Exercise stress testing is reasonable for the evaluation of an asymptomatic young adult with a mean Doppler gradient greater than 40 mm Hg or a peak

Doppler gradient greater than 64 mm Hg or when the patient anticipates athletic participation or pregnancy. (***Level of Evidence: C***)
4. Dobutamine stress testing can be beneficial in the evaluation of a mild aortic valve gradient in the face of low LV ejection fraction and reduced cardiac output. (***Level of Evidence: C***)
5. Magnetic resonance imaging/computed tomography (MRI/CT) can be beneficial to add important information about the anatomy of the thoracic aorta. (***Level of Evidence: C***)
6. Exercise stress testing can be useful to evaluate blood pressure response or elicit exercise-induced symptoms in asymptomatic older adults with AS. (***Level of Evidence: B***)

Class IIb

1. Magnetic resonance angiography may be beneficial in quantifying AR when other data are ambiguous or borderline. (***Level of Evidence: C***)

Class III

1. Exercise stress testing should not be performed in symptomatic patients with AS or those with repolarization abnormality on ECG or systolic dysfunction on echocardiography. (***Level of Evidence: C***)

Management Strategies for Left Ventricular Outflow Tract Obstruction and Associated Lesions

Recommendations for Medical Therapy

Class IIa

1. It is reasonable to treat systemic hypertension in patients with AS while monitoring diastolic blood pressure to avoid reducing coronary perfusion. (***Level of Evidence: C***)
2. It is reasonable to administer beta blockers in patients with bicuspid aortic valve (BAV) and aortic root dilatation. (***Level of Evidence: C***)
3. It is reasonable to use long-term vasodilator therapy in patients with AR and systemic hypertension while carefully monitoring diastolic blood pressure to avoid reducing coronary perfusion. (***Level of Evidence: C***)

Class IIb

1. It may be reasonable to treat patients with BAV and risk factors for atherosclerosis with statins with the aim of slowing down degenerative changes in the aortic valve and preventing atherosclerosis. (***Level of Evidence: C***)

Class III

1. Vasodilator therapy is not indicated for long-term therapy in AR for the following:

a. The asymptomatic patient with only mild to moderate AR and normal LV function. (***Level of Evidence: B***)
b. The asymptomatic patient with LV systolic dysfunction who is otherwise a candidate for AVR. (***Level of Evidence: B***)
c. The asymptomatic patient with either LV systolic function or mild to moderate LV diastolic dysfunction who is otherwise a candidate for AVR. (***Level of Evidence: C***)

Recommendations for Catheter Interventions for Adults With Valvular Aortic Stenosis

Class I

1. In young adults and others without significantly calcified aortic valves and no AR, aortic balloon valvotomy is indicated in the following patients:
    a. Those with symptoms of angina, syncope, dyspnea on exertion, and peak-to-peak gradients at catheterization greater than 50 mm Hg. (***Level of Evidence: C***)
    b. Asymptomatic adolescents or young adults who demonstrate ST or T-wave abnormalities in the left precordial leads on ECG at rest or with exercise and a peak-to-peak catheter gradient greater than 60 mm Hg. (***Level of Evidence: C***)

Class IIa

1. Aortic balloon valvotomy is reasonable in the asymptomatic adolescent or young adult with AS and a peak-to-peak gradient on catheterization greater than 50 mm Hg when the patient is interested in playing competitive sports or becoming pregnant. (***Level of Evidence: C***)

Class IIb

1. Aortic balloon valvotomy may be considered as a bridge to surgery in hemodynamically unstable adults with AS, adults at high risk for AVR, or when AVR cannot be performed secondary to significant comorbidities. (***Level of Evidence: C***)

Class III

1. In older adults, aortic balloon valvotomy is not recommended as an alternative to AVR, although certain younger patients may be an exception and should be referred to a center with experience in aortic balloon valvuloplasties. (***Level of Evidence: B***)
2. In asymptomatic adolescents and young adults, aortic balloon valvotomy should not be performed with a peak-to-peak gradient less than 40 mm Hg without symptoms or ECG changes. (***Level of Evidence: B***)

Recommendations for Aortic Valve Repair/Replacement and Aortic Root Replacement

Class I

1. Aortic valvuloplasty, AVR, or Ross repair is indicated in patients with severe AS or chronic severe AR while they undergo coronary artery bypass grafting, surgery on the aorta, or surgery on other heart valves. (***Level of Evidence: C***)
2. AVR is indicated for patients with severe AS and LV dysfunction (LV ejection fraction less than 50%). (***Level of Evidence: C***)
3. AVR is indicated in adolescents or young adults with severe AR who have:
    a. Development of symptoms. (***Level of Evidence: C***)
    b. Development of persistent LV dysfunction (LV ejection fraction less than 50%) or progressive LV dilatation (LV end-diastolic diameter 4 standard deviations above normal). (***Level of Evidence: C***)
4. Surgery to repair or replace the ascending aorta in a patient with a BAV is recommended when the ascending aorta diameter is 5.0 cm or more or when there is progressive dilatation at a rate greater than or equal to 5 mm per year. (American College of Cardiology et al., 2006) (***Level of Evidence: B***)

Class IIa

1. AVR is reasonable for asymptomatic patients with severe AR and normal systolic function (ejection fraction greater than 50%) but with severe LV dilatation (LV end-diastolic diameter greater than 75 mm or end-systolic dimension greater than 55 mm*). (***Level of Evidence: B***)
2. Surgical aortic valve repair or replacement is reasonable in patients with moderate AS undergoing coronary artery bypass grafting or other cardiac or aortic root surgery. (***Level of Evidence: B***)

*Consider lower threshold values for patients of small stature of either gender.

Class IIb

1. AVR may be considered for asymptomatic patients with any of the following indications:
    a. Severe AS and abnormal response to exercise. (***Level of Evidence: C***)
    b. Evidence of rapid progression of AS or AR. (***Level of Evidence: C***)
    c. Mild AS while undergoing coronary artery bypass grafting or other cardiac surgery and evidence of a calcific aortic valve. (***Level of Evidence: C***)
    d. Extremely severe AS (aortic valve area less than 0.6 cm and/or mean Doppler systolic AV gradient greater than 60 mm Hg) in an otherwise good operative candidate. (***Level of Evidence: C***)
    e. Moderate AR undergoing coronary artery bypass grafting or other cardiac surgery. (***Level of Evidence: C***)
    f. Severe AR with rapidly progressive LV dilation when the degree of LV dilation exceeds an end-diastolic dimension of 70 mm or end-systolic dimension of 50 mm, with declining exercise tolerance, or with abnormal hemodynamic responses to exercise. (***Level of Evidence: C***)
2. Surgical repair may be considered in adults with AS or AR and concomitant ascending aortic dilatation (ascending aorta diameter greater than 4.5 cm) coexisting with AS or AR. (***Level of Evidence: B***)

3. Early surgical repair may be considered in adults with the following indications:
    a. AS and a progressive increase in ascending aortic size. (***Level of Evidence: C***)
    b. Mild AR if valve-sparing aortic root replacement is being considered. (***Level of Evidence: C***)

Class III

1. AVR is not useful for prevention of sudden death in asymptomatic adults with AS who have none of the findings listed under the Class IIa/IIb indications. (***Level of Evidence: B***)
2. AVR is not indicated in asymptomatic patients with AR who have normal LV size and function. (***Level of Evidence: B***)

Recommendations for Key Issues to Evaluate and Follow-Up

Class I

1. Lifelong cardiology follow-up is recommended for all patients with aortic valve disease (AS or AR) (operated or unoperated; refer to "Recommendations for Evaluation of the Unoperated Patient" above). (***Level of Evidence: A***)
2. Serial imaging assessment of aortic root anatomy is recommended for all patients with BAV, regardless of severity. The frequency of imaging would depend on the size of the aorta at initial assessment: if less than 40 mm, it should be reimaged approximately every 2 years; if greater than or equal to 40 mm, it should be reimaged yearly or more often as progression of root dilation warrants or whenever there is a change in clinical symptoms or findings. (***Level of Evidence: B***)
3. Prepregnancy counseling is recommended for women with AS who are contemplating pregnancy. (***Level of Evidence: B***)
4. Patient referral to a pediatric cardiologist experienced in fetal echocardiography is indicated in the second trimester of pregnancy to search for cardiac defects in the fetus. (***Level of Evidence: C***)
5. Women with BAV and ascending aorta diameter greater than 4.5 cm should be counseled about the high risks of pregnancy. (***Level of Evidence: C***)
6. Patients with moderate to severe AS should be counseled against participation in competitive athletics and strenuous isometric exercise. (***Level of Evidence: B***)
7. Echocardiographic screening for the presence of BAV is recommended for first-degree relatives of patients with BAV. (***Level of Evidence: B***)

Isolated Subaortic Stenosis

Management Strategies

Recommendations for Surgical Intervention

Class I

1. Surgical intervention is recommended for patients with subaortic stenosis (SubAS) and a peak instantaneous gradient of 50 mm Hg or a mean gradient of 30 mm Hg on echocardiography-Doppler. (***Level of Evidence: C***)
2. Surgical intervention is recommended for SubAS with less than a 50-mm Hg peak or less than a 30-mm Hg mean gradient and progressive AR and an LV dimension at end-systolic diameter of 50 mm or more or LV ejection fraction less than 55%. (***Level of Evidence: C***)

Class IIb

1. Surgical resection may be considered in patients with a mean gradient of 30 mm Hg, but careful follow-up is required to detect progression of stenosis or AR. (***Level of Evidence: C***)
2. Surgical resection may be considered for patients with less than a 50-mm Hg peak gradient or less than a 30-mm Hg mean gradient in the following situations:
    a. When LV hypertrophy is present. (***Level of Evidence: C***)
    b. When pregnancy is being planned. (***Level of Evidence: C***)
    c. When the patient plans to engage in strenuous/competitive sports. (***Level of Evidence: C***)

Class III

1. Surgical intervention is not recommended to prevent AR for patients with SubAS if the patient has trivial left ventricular outflow tract (LVOT) obstruction or trivial to mild AR. (***Level of Evidence: C***)

Recommendations for Key Issues to Evaluate and Follow-Up

Class I

1. Lifelong cardiology follow-up, including evaluation by and/or consultation with a cardiologist with expertise in ACHD, is recommended for all patients with SubAS, repaired or not. (***Level of Evidence: C***)
2. The unoperated asymptomatic adult with stable LVOT obstruction due to SubAS and a mean gradient less than 30 mm Hg without LV hypertrophy or significant AR should be monitored at yearly intervals for increasing obstruction, the development or progression of AR, and the evaluation of systolic and diastolic LV function. (***Level of Evidence: B***)

Class IIa

1. Stress testing to determine exercise capability, symptoms, ECG changes or arrhythmias, or increase in LVOT gradient is reasonable in the presence of otherwise equivocal indications for intervention. (***Level of Evidence: C***)

Recommendations for Evaluation of the Unoperated Patient

Class I

1. Transthoracic echocardiography (TTE) and/or transesophageal echocardiography (TEE) with Doppler and either MRI or CT should be performed to assess the anatomy of the LVOT, the ascending aorta, coronary artery anatomy and flow, and main and branch pulmonary artery anatomy and flow. (***Level of Evidence: C***)
2. Assessment of anatomy and flow in the proximal renal arteries is recommended in ACHD patients with SupraAS. (***Level of Evidence: C***)
3. Assessment of systolic and diastolic ventricular function is recommended in ACHD patients with SupraAS. (***Level of Evidence: C***)
4. Assessment of aortic and mitral valve anatomy and function is recommended in ACHD patients with SupraAS. (***Level of Evidence: C***)
5. Adults with a history or presence of SupraAS should be screened periodically for myocardial ischemia. (***Level of Evidence: C***)

Class IIa

1. Exercise testing, dobutamine stress testing, positron emission tomography, or stress sestamibi with adenosine studies can be useful to evaluate the adequacy of myocardial perfusion. (***Level of Evidence: C***)

Management Strategies for Supravalvular Left Ventricular Outflow Tract (LVOT)

Recommendations for Interventional and Surgical Therapy

Class I

1. Operative intervention should be performed for patients with supravalvular LVOT obstruction (discrete or diffuse) with symptoms (i.e., angina, dyspnea, or syncope) and/or mean gradient greater than 50 mm Hg or peak instantaneous gradient by Doppler echocardiography greater than 70 mm Hg. (***Level of Evidence: B***)
2. Surgical repair is recommended for adults with lesser degrees of supravalvular LVOT obstruction and the following indications:
    a. Symptoms (i.e., angina, dyspnea, or syncope). (***Level of Evidence: B***)
    b. LV hypertrophy. (***Level of Evidence: C***)
    c. Desire for greater degrees of exercise or a planned pregnancy. (***Level of Evidence: C***)
    d. LV systolic dysfunction. (***Level of Evidence: C***)
3. Interventions for coronary artery obstruction in patients with SupraAS should be performed in ACHD centers with demonstrated expertise in the interventional management of such patients. (***Level of Evidence: C***)

Recommendations for Key Issues to Evaluate and Follow-Up

Class I

1. Both operated and unoperated patients with SupraAS should be followed up annually at a regional ACHD center. (***Level of Evidence: C***)

2. Long-term psychosocial assessment and oversight, including the need for legal guardianship, are recommended for patients with Williams syndrome. (***Level of Evidence: C***)

Recommendations for Reproduction

Class I

1. SupraAS, whether associated with Williams syndrome or nonsyndromic, has a strong likelihood of being an inherited disorder. Undetected family members may be at risk for hypertension, coronary disease, or stroke; therefore, all available relatives should be screened. (***Level of Evidence: C***)
2. Patients with SupraAS and significant obstruction, coronary involvement, or aortic disease should be counseled against pregnancy. (***Level of Evidence: C***)

Aortic Coarctation

Recommendations for Clinical Evaluation and Follow-Up

Class I

1. Every patient with systemic arterial hypertension should have the brachial and femoral pulses palpated simultaneously to assess timing and amplitude evaluation to search for the "brachial-femoral delay" of significant aortic coarctation. Supine bilateral arm (brachial artery) blood pressures and prone right or left supine leg (popliteal artery) blood pressures should be measured to search for differential pressure. (***Level of Evidence: C***)
2. Initial imaging and hemodynamic evaluation by TTE, including suprasternal notch acoustic windows, is useful in suspected aortic coarctation. (***Level of Evidence: B***)
3. Every patient with coarctation (repaired or not) should have at least 1 cardiovascular MRI or CT scan for complete evaluation of the thoracic aorta and intracranial vessels. (***Level of Evidence: B***)

Management Strategies for Coarctation of the Aorta

Recommendations for Interventional and Surgical Treatment of Coarctation of the Aorta in Adults

Class I

1. Intervention for coarctation is recommended in the following circumstances:
    a. Peak-to-peak coarctation gradient greater than or equal to 20 mm Hg. (***Level of Evidence: C***)
    b. Peak-to-peak coarctation gradient less than 20 mm Hg in the presence of anatomic imaging evidence of significant coarctation with radiological evidence of significant collateral flow. (***Level of Evidence: C***)
2. Choice of percutaneous catheter intervention versus surgical repair of native discrete coarctation should be determined by consultation with a team of

ACHD cardiologists, interventionalists, and surgeons at an ACHD center. (***Level of Evidence: C***)
3. Percutaneous catheter intervention is indicated for recurrent, discrete coarctation and a peak-to-peak gradient of at least 20 mm Hg. (***Level of Evidence: B***)
4. Surgeons with training and expertise in CHD should perform operations for previously repaired coarctation and the following indications:
    a. Long recoarctation segment. (***Level of Evidence: B***)
    b. Concomitant hypoplasia of the aortic arch. (***Level of Evidence: B***)

Class IIb

1. Stent placement for long-segment coarctation may be considered, but the usefulness is not well established, and the long-term efficacy and safety are unknown. (***Level of Evidence: C***)

Recommendations for Key Issues to Evaluate and Follow-Up

Class I

1. Lifelong cardiology follow-up is recommended for all patients with aortic coarctation (repaired or not), including an evaluation by or consultation with a cardiologist with expertise in ACHD. (***Level of Evidence: C***)
2. Patients who have had surgical repair of coarctation at the aorta or percutaneous intervention for coarctation of the aorta should have at least yearly follow-up. (***Level of Evidence: C***)
3. Even if the coarctation repair appears to be satisfactory, late postoperative thoracic aortic imaging should be performed to assess for aortic dilatation or aneurysm formation. (***Level of Evidence: B***)
4. Patients should be observed closely for the appearance or reappearance of resting or exercise-induced systemic arterial hypertension, which should be treated aggressively after recoarctation is excluded. (***Level of Evidence: B***)
5. Evaluation of the coarctation repair site by MRI/CT should be performed at intervals of 5 years or less, depending on the specific anatomic findings before and after repair. (***Level of Evidence: C***)

Class IIb

1. Routine exercise testing may be performed at intervals determined by consultation with the regional ACHD center. (***Level of Evidence: C***)

Definitions:

Applying Classification of Recommendations and Level of Evidence

Â	**SIZE OF TREATMENT EFFEC**		
Â	**CLASS I** *Benefit >>> Risk*	**CLASS IIa** *Benefit >> Risk* *Additional studies with*	**CLASS IIb** *Benefit ≥ Risk* *Additional stud*

Â		SIZE OF TREATMENT EFFEC		
		Procedure/Treatment **SHOULD** be performed/administered	*focused objectives needed* **IT IS REASONABLE** to perform procedure/administer treatment	*objectives nee registry data w helpful* Procedure/Trea **MAY BE CONS**
Estimate of Certainty (Precision) of Treatment Effect	**LEVEL A** Multiple population evaluated* Data derived from multiple randomized clinical trials or meta-analyses	• Recommendation that procedure or treatment is useful/effective • Sufficient evidence from multiple randomized trials or meta-analyses	• Recommendation in favor of treatment of procedure being useful/effective • Some conflicting evidence from multiple randomized trials or meta-analyses	• Recomm usefuln less we • Greater evidenc multiple trials or analyse
	LEVEL B Limited population evaluated* Data derived from a single randomized clinical trial or nonrandomized studies	• Recommendation that procedure or treatment is useful/effective • Evidence from single randomized trial or nonrandomized studies	• Recommendation in favor of treatment of procedure being useful/effective • Some conflicting evidence from single randomized trial or nonrandomized studies	• Recomm usefuln less we • Greater evidenc random nonran studies
	LEVEL C Very limited population evaluated* Only consensus opinion of experts, case studies or standard of care.	• Recommendation that procedure or treatment is useful/effective • Only expert opinion, case studies, or standard-of-care	• Recommendation in favor of treatment of procedure being useful/effective • Only diverging expert opinion, case studies, or standard-of-care	• Recomm usefuln less we • Only di opinion or stan

*Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as gender, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use. A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Even though randomized trials are not available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

Note: In 2003, the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines developed a list of suggested phrases to use when writing recommendations. All guideline recommendations have been written in full sentences that express a complete thought, such that a recommendation, even if separated and presented apart from the rest of the document (including headings above sets of recommendations), would still convey the full intent of the recommendation. It is hoped that this will increase readers' comprehension of the guidelines and will allow queries at the individual recommendation level. (See Table 1 in the original guideline document for a list of suggested phrases for writing recommendations.)

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

REFERENCES SUPPORTING THE RECOMMENDATIONS

References open in a new window

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of supporting evidence is identified and graded for each recommendation (see "Major Recommendations").

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Appropriate management of patients with adult congenital heart disease and left-sided heart obstructive lesions

POTENTIAL HARMS

Complications of Medical Therapy

Judicious afterload reduction in patients with hypertension to reduce systolic blood pressure and lower left ventricular (LV) wall tension may delay onset of LV dilatation or dysfunction but should be balanced against the risk of reducing diastolic coronary perfusion.

Complications of Surgical Interventions

- Prosthetic valve complications include endocarditis, thrombosis, periprosthetic regurgitation with or without hemolysis, and obstruction related to pannus in growth. Patients who undergo the Ross procedure (placement of the native pulmonary valve in the aortic position and pulmonary or aortic homograft replacement of the pulmonary valve) are at risk of developing autograft dilatation with progressive neo-aortic regurgitation (AR), right-sided pulmonary homograft obstruction and/or regurgitation, and occasionally myocardial ischemia and/or infarct related to proximal coronary artery obstruction or kinking. Patients who undergo the Bentall procedure (aortic

root replacement with a composite valve and graft with coronary reimplantation) are also at risk for proximal coronary obstruction.
- Potential operative complications of surgical repair of subaortic stenosis (SubAS) include injury to the aortic or mitral valves, complete heart block, or creation of a ventricular septal defect (VSD). Postoperative complications may include damage to the aortic or mitral valve, heart block, iatrogenic VSD, and infective endocarditis.
- Morbidity in adults with reoperation for coarctation can be considerable and may include significant early postoperative bleeding, pleural effusion, lung contusion, recurrent laryngeal nerve palsy, or phrenic nerve injury (with hemidiaphragmatic paresis or paralysis). Other postoperative complications include recoarctation and hypertension. Aneurysm formation at the repair site can occur after patch aortoplasty (particularly with the use of a Dacron patch) or resection of the coarctation shelf. False aneurysms may also occur at the repair site. Late dissection proximal or distal to the repair site can occur. Paraplegia secondary to spinal cord ischemia is rare but is more common with poor collateral circulation. Arm claudication or subclavian steal syndrome is rare but in particular may occur after use of the subclavian flap technique.

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

- These practice guidelines are intended to assist healthcare providers in clinical decision making by describing a range of generally acceptable approaches for diagnosis, management, and prevention of specific diseases or conditions. Clinicians should consider the quality and availability of expertise in the area where care is provided. These guidelines attempt to define practices that meet the needs of most patients in most circumstances. The recommendations reflect a consensus of expert opinion after a thorough review of the available current scientific evidence and are intended to improve patient care.
- Patient adherence to prescribed and agreed upon medical regimens and lifestyles is an important aspect of treatment. Prescribed courses of treatment in accordance with these recommendations are only effective if they are followed. Because lack of patient understanding and adherence may adversely affect outcomes, physicians and other healthcare providers should make every effort to engage the patient's active participation in prescribed medical regimens and lifestyles.
- If these guidelines are used as the basis for regulatory or payer decisions, the goal is quality of care and serving the patient's best interest. The ultimate judgment regarding care of a particular patient must be made by the healthcare provider and the patient in light of all of the circumstances presented by that patient. There are circumstances in which deviations from these guidelines are appropriate.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

IMPLEMENTATION TOOLS

Slide Presentation

For information about availability, see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Living with Illness

IOM DOMAIN

Effectiveness
Patient-centeredness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, del Nido P, Fasules JW, Graham TP, Hijazi ZM, Hunt SA, King ME, Landzberg MJ, Miner PD, Radford MJ, Walsh EP, Webb GD, Smith SC Jr, Jacobs AK, Adams CD, Anderson JL, Antman EM, Buller CD, Creager MA, Ettinger SM, Halperin JL, Hunt SA, Krumholz HM, Kushner FG, Lytle BW, Nishimura RA, Page RL, Riegel B, Tarkington LG, Yancy CW. Left-sided heart obstructive lesions: aortic valve disease, subvalvular and supravalvular aortic stenosis, associated disorders of the ascending aorta, and coarctation. In: ACC/AHA 2008 guidelines for the management of adults [trunc]. J Am Coll Cardiol 2008;52(23):e185-97.

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2008

GUIDELINE DEVELOPER(S)

American College of Cardiology Foundation - Medical Specialty Society
American Heart Association - Professional Association

SOURCE(S) OF FUNDING

The American College of Cardiology Foundation and the American Heart Association. No outside funding accepted.

GUIDELINE COMMITTEE

American College of Cardiology/American Heart Association Task Force on Practice Guidelines

Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease

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†International Society for Adult Congenital Heart Disease representative.

‡Society for Cardiovascular Angiography and Interventions representative.

§American Society of Echocardiography representative.

||Heart Rhythm Society representative.

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#Former Task Force member during this writing effort.

**Immediate past chair.

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

The American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines makes every effort to avoid actual, potential, or perceived conflicts of interest that might arise as a result of industry relationships or personal interests among the writing committee. Specifically, all members of the writing committee, as well as peer reviewers of the document, are asked to disclose all such relationships that might be perceived as real or potential conflicts of interest. Writing committee members are also strongly encouraged to declare previous relationships with industry that might be perceived as relevant to guideline development. If a writing committee member develops a new relationship with industry during their tenure, they are required to notify guideline staff in writing. These statements are reviewed by the parent task force, reported orally to all members at each meeting of the writing committee, and updated and reviewed by the writing committee as changes occur.

Author Relationships With Industry and Other Entities–ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

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This table represents the relevant relationships of committee members with industry and other entities that were reported orally at the initial writing committee meeting and updated in conjunction with all meetings and conference calls of the writing committee during the document development process. It does not necessarily reflect relationships with industry at the time of publication. A person is deemed to have a significant interest in a business if the interest represents ownership of 5% or more of the voting stock or share of the business entity, or ownership of $10,000 or more of the fair market value of the business entity; or if funds received by the person from the business entity exceed 5% of the person's gross income for the previous year. A relationship is considered to be modest if it is less than significant under the preceding definition. Relationships in this table are modest unless otherwise noted.

See Appendix 2 in the original guideline document for peer reviewer relationships with industry.

ENDORSER(S)

American Society of Echocardiography - Professional Association
Heart Rhythm Society - Professional Association
International Society for Adult Congenital Heart Disease - Disease Specific Society

Society for Cardiovascular Angiography and Interventions - Medical Specialty Society
Society of Thoracic Surgeons - Medical Specialty Society

GUIDELINE STATUS

This is the current release of the guideline.

The guidelines will be reviewed annually by the American College of Cardiology/American Heart Association (ACC/AHA) Task Force on Practice Guidelines and considered current unless they are updated, revised, or withdrawn from distribution.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the American College of Cardiology (ACC) Web site; electronic copies are also available in PDF from the American Heart Association (AHA) Web site.

Print copies: Available from the American College of Cardiology, Resource Center, 9111 Old Georgetown Rd, Bethesda, MD 20814-1699; (800) 253-4636 (US only).

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

- ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: executive summary. A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease). J Am Coll Cardiol, 2008; 52:1890-1947. Electronic copies: Available from the American College of Cardiology (ACC) Web site. Also available in Portable Document Format (PDF) from the American Heart Association (AHA) Web site.
- ACC/AHA 2008 guidelines for the management of adults with congenital heart disease. Slide set. 2008. 88 p. Electronic copies: Available from the American College of Cardiology (ACC) Web site.
- Methodology manual for ACC/AHA Guideline Writing Committees. Methodologies and policies from the ACC/AHA Task Force on Practice Guidelines. 2006 Jun. 61 p. Electronic copies: Available in PDF from the American College of Cardiology (ACC) Web site.

Print copies: Available from the American College of Cardiology, 9111 Old Georgetown Road, Bethesda, Maryland 20814-1699.

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI Institute on September 18, 2009.

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